Sinonasal Glomangiopericytoma : A Case Report

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－ ABSTRACT －

Glomangiopericytoma (sinonasal-type hemangiopericytoma) is a rare sinonasal neoplasm, arising from pericytes surrounding capillaries. This tumor accounts for less than 0.5% of all sinonasal tumors. It is a borderline or low grade malignant tumor and a radial surgical resection is considered. The endoscopic and imaging findings are nonspecific. The final diagnosis is based on histopathology and immunochemistry. A 71-year-old woman presented with glomangiopericytoma of the nasal cavity manifestating nasal obstruction and epistaxis. Complete resection of the mass was performed endoscopically. We report herein a case of sinonasal glomangiopericytoma. **(J Clinical Otolaryngol 2015;26:87-91)**

KEY WORDS : Nasal cavity · Hemangiopericytoma · Epistaxis.

Introduction

Glomangiopericytoma (sinonasal-type hemangiopericytoma) is an unusual vascular tumor. This tumor originate in extravascular cells called pericytes, which presumably are modified contractile smooth-muscle cells found on the external surface of capillaries and post-capillary venules.[1)] The World Health Organization (WHO) classified glomangiopericytoma in 2005. This classification includes the tumor described as sinonasal type hemangiopericrytoma, hemangiopericytoma-like tumor and sinonasal glomus tumour.[2)] This tumor differs from conventional soft tissue hemangiopericytoma in location, biologic behavior, and histologic features. In national report, Byun JY, et al first report about a case of glomangiopericytoma of the ethmoid sinus not the term of hemangiopericytoma.[3)]

Here, we report a case of 71-year-old woman with sinonasal glomangiopericytoma in terms of clinicopathologic and imaging findings.

Case Report

A 71-year-old woman presented with a 3-month history of nasal obstruction and frequent epistaxis. Nasal endoscopy showed a reddish polyp occupying the left posterior nasal cavity (Fig. 1). Computed tomography (CT) showed an approximately 2-cm soft tissue mass with heterogeneous contrast enhancement in the left nasal cavity arising from the posterior nasal septum. There was no bony destruction or invasion of the surrounding tissue (Fig. 2). Magenetic resonance imaging (MRI) confirmed a circumscribed lobular soft tissue mass in the left nasal cavity. The mass showed iso-intense signal on T1-weighted images along with strong contrast enhancement and hyper intense signal on T2-weighted images. There were no signal voids in vascular signal flow within the mass or enlarged vas-

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cular structures as tumor feeders around the mass (Fig. 3). The radiologic diagnosis of the nasal mass was a benign solid tumor, such as angiomatous polyp, hemangioma or inverted papilloma. Endoscopic biopsy of the nasal mass was performed. There was no significant bleeding after biopsy. We performed endoscopic excision of the nasal mass. The safety margin including 2 mm normal nasal mucosa was indicated by needle type monopolar cautery tip. Subperiosteal and subperichondrial dissection around mass were done by Freer elevator and curette. The tumor was completely removed, the excised specimen contained soft polypoid grayish pink mass with smooth surface. After tumor removal, for preventing recurrence, vomer was partially cut by back bite forceps and protruded hard palate was drilled out by sinur burr.

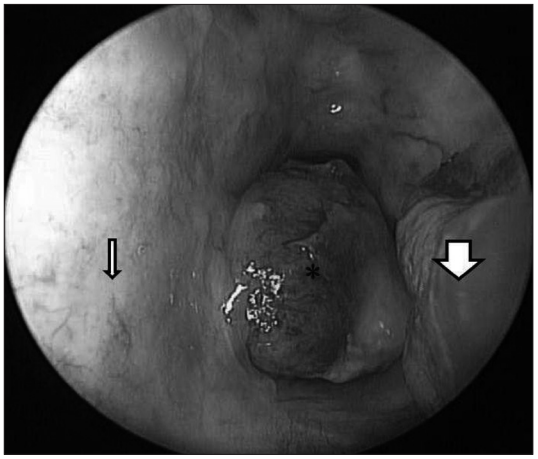

Fig. 1. A 71-year-old woman with glomangiopericytoma. Nasal endoscopy showed a reddish polyp (asterisk) occupying the left posterior nasal cavity. Nasal septum (narrow black arrow) and left inferior turbinate (wide black arrow).

Histologically, the mass was covered with intact respiratory epithelium with partial surface erosion. The neoplastic cells were mainly distributed in the perivascular area showing diffuse, syncytial growth pattern. The tumor cells demonstrated plump eosinophilic cytoplasm with vesicular nuclei and small, prominent nucleoli. Although mild pleomorphism was seen, significantly atypical features were not evident. A few mitotic figures (<1 mitoses/10 high-power fields) were identified and no necrosis was found (Fig. 4A, B, C). Immunohistochemically, the neoplastic cells were diffusely positive for vimentin and focally positive for smooth muscle actin, revealing the myoid phenotype (Fig. 4D). CD34 staining was negative, which excluded the possibility of vascular tumors or solitary fibrous tumors. Interestingly, the neoplastic cells showed a positive reaction for cytokeratin, which is a immunohistochemical marker to detect epithelial cell origin (Fig. 4E). However, all epithelial tumors were exclud-

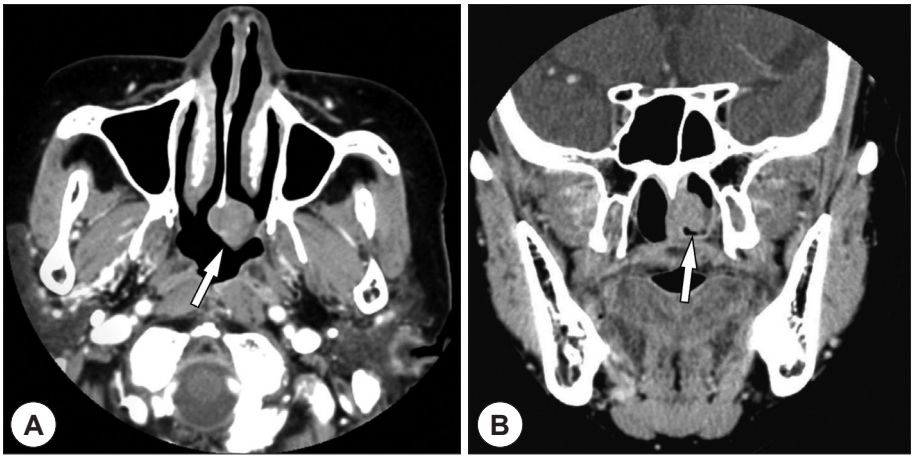

Fig. 2. Axial (A) and coronal (B) computed tomography (CT) with contrast enhancement show a 2-cm soft tissue mass (arrow) with heterogeneous contrast enhancement in the left nasal cavity arising from the posterior nasal septum. There is no bony destruction or invasion of the surrounding tissue.

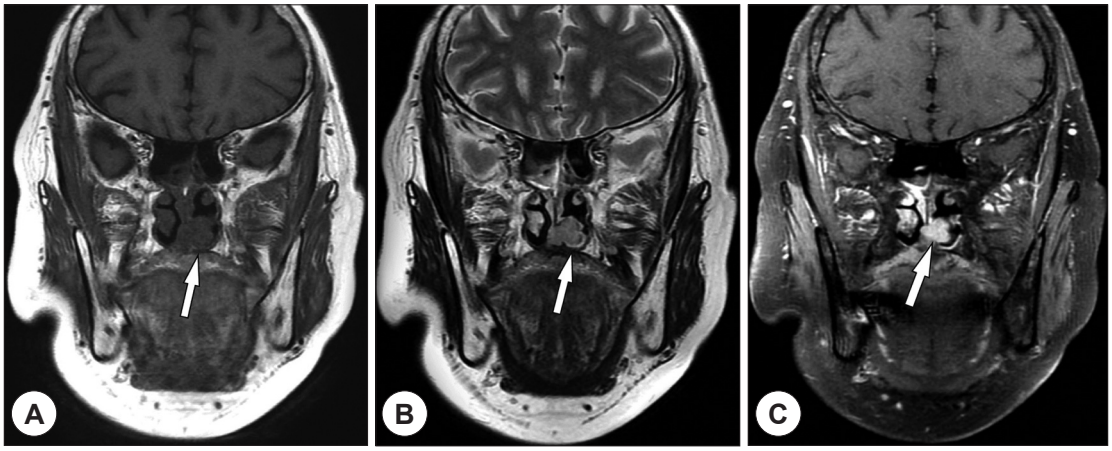

Fig. 3. Magnetic resonance imaging (MRI) of the nasal cavity. The mass (arrow) shows iso-intense signal on a coronal T1-weighted image (A), a hyper intense signal on a T2-weighted image (B) and strong contrast enhancement on an enhanced T1-weighted image (C). There are no signal voids in vascular signal flow within the mass or, enlarged vascular structures as tumor feeder around the mass.

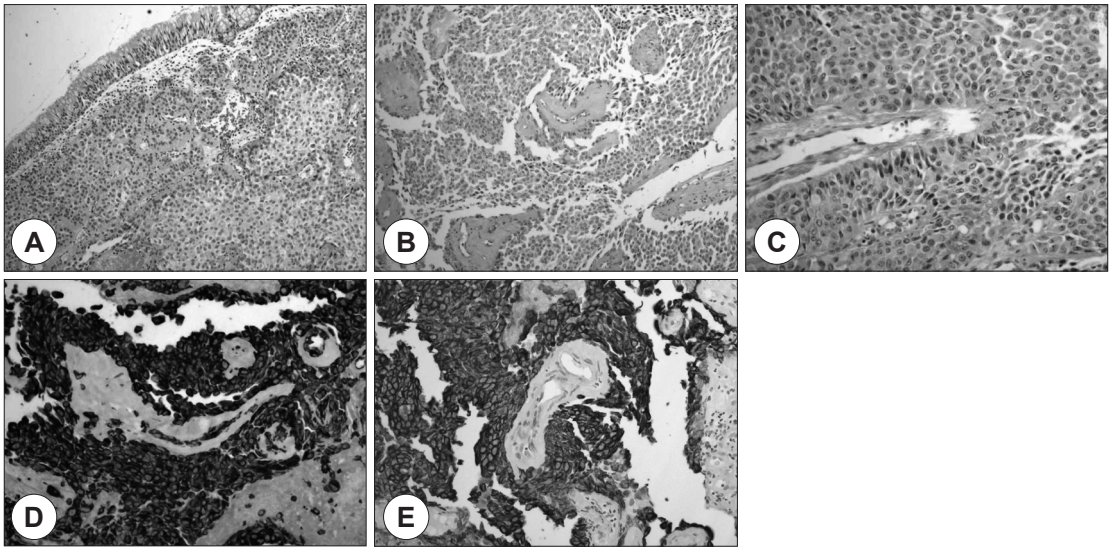

Fig. 4. The tumor cells show intact respiratory surface epithelium overlying neoplastic cell nests (A) and are arranged in a diffuse syncytial pattern with perivascular hyalinization (B, hematoxylin & eosin stain, ×200). A high-power microscopic view shows perivascular proliferation of tumor cells with mild pleomorphism (C, hematoxylin & eosin stain, ×400). Immunohistochemical staining for vimentin highlights the perivascular neoplastic cell population (D, ×400). The tumor cells express pancytokeratin in their cytoplasm (E, ×400).

ed in the differential diagnoses, based on the growth pattern of tumor cells and immunohistochemical expression of mesenchymal markers. Based on these pathological findings, the patient was diagnosed with glomangiopericytoma.

The patient's postoperative course was uneventful. There was no recurrence, symptom at 12 months follow up.

Discussion

Conventional soft tissue hemangiopericytoma ac-

count for 2% to 3% of all soft tissue sarcomas and 1% of all vascular tumors.[1,4] This tumor commonly arises in the musculoskeletal system, skin, and retroperitonum. Approximately 15% to 30 % of all hemangiopericytomas occur in the head and neck area; only 5% occur in the sinonasal area.[1,5] Sinonasal-type hemangiopericytoma is now termed glomangiopericytoma as a separated lesion.[6] Glomangiopericytoma is believed to behave less aggressively than soft tissue hemangiopericytoma which occurs in other parts of the body and is within the category of borderline and low-grade malignant potential soft tissue tumors. Local recurrence is seen 25% of all glomangiopericytoma cases, and metastasis occurs in 5% of all cases of sinonasal type, as compared to 25−60% in somatic hemangiopericytoma.[7-9] The 5-year survival rate of patients with glomangiopericytoma is approximately 88%.[1]

Males and females are equally affected, but some studies report an equal-to-slight female predominance.[1,10,11] Most cases occurs in patients' third and fifth decades of life without racial predominance.[4] However there have been reports of a slight increase in sixth and seventh decades.[4,11] The most presenting symptoms are epistaxis and nasal obstruction.[1,11] Rarely oncogenic osteomalacia is caused by glomangiopericytoma, manifesting as symptoms characteristic of hypophosphatemic osteromalacia, such as recurrent fractures, bone pain, and gait disturbance.[12] Compression symptoms such as proptosis, epiphora, and diplopia are rare but can be found with increasing tumor size or intraorbital extension. Endoscopically, glomangiopericytoma is soft and tan-colored, and is typically enlarged slowly over a period of months or years.[1] Although its etiology is unclear, it has been frequently associated with trauma, long-term steroid use, pregnancy, hypertension and hormone imbalance.[10]

CT of glomangiopericytoma demonstrates tumor involvement of soft tissues of the nasal cavity and paranasal sinuses that can be enhanced after contrast administration. On MR imaging, glomangiopericytoma appears as a solid mass with iso-intense signals on T1-weighted images with diffuse enhancement after intravenous administration of gadolinium. On T2-weighted images, glomangiopericytoma shows isointense to hyper intense signals. However the hyper intense signals differ from that of inflammatory fluid caused by sinus obstruction. In highly vascular tumors, signal voids may also be seen on MRI. Radiologic findings of glomangiopericytoma are similar to those of common benign solid tumors such as inverted papillomas, angiomatous polyps, inflammatory polyps. However, differential diagnosis of glomangiopericytoma from such tumors by using preoperative biopsy is essential, because misdiagnosis may result in an incomplete excision. This is an important risk factor for tumor recurrence. In addition, misdiagnosis may also lead to significant blood loss during and after surgery.[13]

The histologic findings of glomangiopericytoma are spindle cell neoplasia with hemangiopericytoma-like vessles and perivascular hyalinization of capillary-sized vessels. Glomangiopericytoma is characterized by diffuse growth of closely packed cells, formation of short fascicles, storiform, whorled, or palisaded patterns, and being interspersed with numerous thin-walled branching staghorn vessels. In immunohistochemical staining, this tumor is positive for actin and vimentin and negative for cytokeratins.[5,7,14] Cytokeratins are proteins of keratin-containing intermediate filaments and commonly used as epithelial markers. There have been many reports showing that cytokeratin can be expressed in tumors of non-epithelial origin.[7] However, glomangiopericytoma positive for cytokeratin has never been reported. We present the first case of glomangiopericytoma aberrantly expressing pancytokeratin. The reason for the positive immunoreaction for pancytokeratin is unclear. The differential diagnosis of glomangiopericytoma includes a variety of spindle cell and vascular neoplasms, such as lobular capillary hemangioma, solitary fibrous tumor, and angiofibroma.

Sinonasal tumors can be successfully removed through open or endoscopic approaches.[8,15] Glomangiopericytoma is categorized as a borderline low-grade ma-

lignant tumor by the WHO classification. Local recurrence and distant metastasis occur in about 25% and 5% of all cases, respectively. Wide local excision of glomangiopericytoma is the treatment of choice. However, endoscopic resection is adequate when the mass is small and confined to its site of origin without extension to surrounding tissue.[1)] Postoperative adjuvant therapy is limitedly used, when a mass is incompletely resected due to greater tumor size and infiltration into surrounding tissue. Incomplete resection may lead to tumor recurrence as residual tumor.

In summery, glomangiopericytoma is a rare sinonasal tumor. However, it should be considered in the differential diagnosis of sinonasal solitary tumor. Preoperative radiologic and histologic evaluations of size, location, characteristics, and type are important because glomangiopericytoma is regarded as a borderline low-grade malignancy.

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